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Gene Section
Mini Review

NUP98 (nucleoporin 98 kDa)

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Identity

HGNC (Hugo): NUP98

Location: 11p15

DNA/RNA

Transcription

3.6, 6.5, 7.0 kb mRNA.

Protein

Description

920 amino acids; 97 kDa; contains repeated motifs (GLFG and FG) in N-term and a RNA binding motif in C-term.

Expression

Wide.

Localisation

Nuclear membrane localisation.

Function

Nucleoporin: associated with the nuclear pore complex; role in nucleocytoplasmic transport processes.

Homology

Member of the GLFG nucleoporins.

Implicated in

t(7;11)(p15;p15)/ANLL --> NUP98/HOXA9

Disease
M2-M4 ANLL mostly; occasionally: CML-like cases.

Prognosis
Mean survival: 15 months.

Cytogenetics
Sole anomaly most often.

Hybrid/Mutated gene
5' NUP98 - 3' HOXA9.

Abnormal protein
Fuses the GLFG repeat domains of NUP98 to the HOXA9 homeobox.

inv (11)(p15q22)/MDS or ANLL --> NUP98/DDX10

Disease
Therapy related MDS and ANLL; de novo ANLL.

Hybrid/Mutated gene
5' NUP98 - 3' DDX10.

Abnormal protein
Fuses the GLFG repeat domains of NUP98 to the acidic domain of DDX11.

t(1;11)(q23;p15) --> NUP98/PMX1

t(2;11)(q31;p15)/treatment related leukaemia --> NUP98/HOXD13

Disease
So far, only 1 case of treatment related myelodysplasia evolving towards M6 acute non lymphocytic leukaemia.

Hybrid/Mutated gene

5' NUP98 - 3' HOXD13.

Abnormal protein

Fuses the GLFG repeat domains of NUP98 to the HOXD13 homeodomain.

t(11;12)(p15;q13)/treatment related leukemia

Hybrid/Mutated gene

5' NUP98 - 3' unknown.

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